etin level. Other explanations include intermittent production or the release of erythropoietin[13] or a circadian variation of the erythropoietin level.[19]

Cerebellar hemangioblastomas are benign tumors that can be solid, cystic, or a combination of both. Although they are benign, their course might be malignant because of their tendency to recur as long as ten years after initial tumor excision. The frequency of recurrence is approximately 25%.[1,3,20-22] When the clinical profiles and histopathologic features of 26 patients with recurring and nonrecurring hemangioblastomas were retrospectively examined, patients with recurring tumors tended to be younger, with a mean age of 22.3 versus 43.8 years for nonrecurring tumors; they had a shorter duration of symptoms, a smaller volume of tumor, a higher hematocrit, and the tumors were multicentric.[20] In addition, patients with von Hippel-Lindau disease were prone to recurrent tumors. Men had a higher incidence of tumor recurrence than women. Pathologically recurring tumors were small and noncystic, whereas nonrecurring tumors were larger and highly cystic. In addition, incomplete surgical excision correlated with tumor recurrence.[22]

Total excision is curative. Radiation therapy with 40 to 50 Gy (4,000 to 5,500 rad) over four to six weeks is recommended for tumors not amenable to complete excision, like solid hemangioblastomas that commonly occur in the brain stem. Patients receiving 40 to 50 Gy had a sevenfold higher chance of 15-year survival than patients receiving low-dose radiotherapy of 20 to 30 Gy.[23]

In summary, patients who present with secondary erythrocytosis and neurologic findings should be suspected to have central nervous system hemangioblastomas. Patients with von Hippel-Lindau disease and their families should be counseled because it is an autosomal dominant disorder that carries a poor prognosis if it goes unrecognized.

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Pulmonary Hemorrhage in an Adolescent With Henoch-Schönlein Purpura

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HENOCH-SCHÖNLEIN purpura is a vasculitic syndrome, primarily observed in children, that is characterized by a nonthrombocytopenic purpuric rash, arthralgia or arthritis, abdominal pain often with gastrointestinal bleeding, and nephritis. A high frequency of decreased diffusion capacity in children has been demonstrated with Henoch-Schönlein purpura,[1] but clinically important pulmonary disease is unusual. We report the case of a 15-year-old boy with Henoch-Schönlein purpura in whom acute, bilateral, life-threatening pulmonary hemorrhage developed.

Report of a Case

The patient, a previously healthy 15-year-old boy, was admitted to a local hospital because for two days he had had fever, sore throat, arthralgia, abdominal pain, and an urticarial or purpuric rash on his extremities and buttocks. He was diagnosed with streptococcal pharyngitis by throat culture and was treated with penicillin. His blood pressure progressively increased, and he had heme-positive stools, hematuria, and proteinuria. The rash pro-

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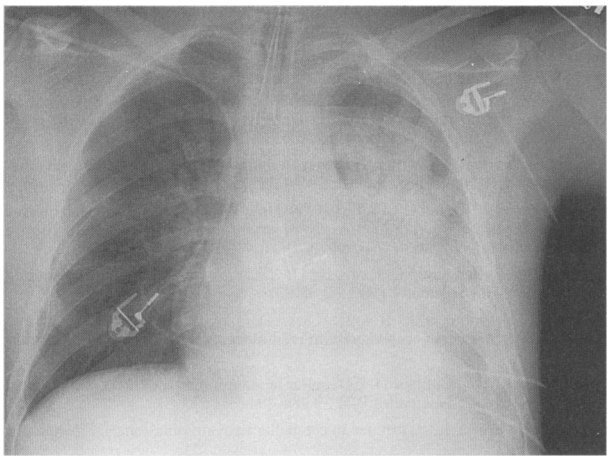

Figure 1.—A chest radiograph was taken about 3 hours after the patient's acute respiratory decompensation. A large right pleural effusion has resolved with the placement of a chest tube. There are new, diffuse alveolar infiltrates with air bronchograms throughout the left lung fields.

gressed to large, palpable purpuric lesions on the distal upper extremities, thighs, and buttocks, and he was diagnosed with Henoch-Schönlein purpura.

Ten days after his symptoms began, the patient was transferred to Madigan Army Medical Center (Tacoma, Washington) where the diagnosis was confirmed by the presence of a palpable purpuric rash primarily on the lower extremities and buttocks, severe arthralgia and arthritis, abdominal pain associated with grossly bloody stools, and nephritis. His blood pressure was 172/104 mm of mercury despite treatment with nifedipine, hydralazine hydrochloride, and furosemide. Laboratory tests elicited the following values: leukocyte count, 36.5 × 10^9 per liter (36,500 per mm^3), with a predominance of neutrophils; hematocrit, 0.39 (39%); platelet count, 313 × 10^9 per liter (313,000 per mm^3); erythrocyte sedimentation rate, 7 mm per hour; serum urea nitrogen, 25.5 mmol per liter (71 mg per dl); serum creatinine, 115 μmol per liter (1.3 mg per dl); serum albumin, 18 grams per liter; C3, 0.6 grams per liter (normal, 0.6 to 1.2), and C4, 0.15 grams per liter (normal, 0.2 to 0.5); and a negative antinuclear antibody (ANA) titer.

Two days after admission, a right pleural effusion developed, and a thoracentesis was done, removing 300 ml of pleural fluid—a transudate, with a ratio of pleural fluid lactate dehydrogenase (LDH) to serum LDH of 0.07. About an hour later, the patient became acutely dyspneic, and his oxygen saturation decreased abruptly to 80% while he was receiving 100% oxygen. An endotracheal tube was inserted, and large amounts of red frothy secretions were suctioned from the trachea. Mechanical ventilation was begun and a right chest tube placed; the pleural fluid evacuated by the chest tube was not grossly bloody (pleural fluid erythrocyte count, 2.44 × 10^9 per liter [2,440 per mm^3]). A chest radiograph revealed new infiltrates in the left upper and lower lung fields, no pneumothorax, and resolution of the right pleural effusion (Figure 1). His hematocrit decreased from 0.32 to 0.22, and he was transfused with four units of packed red blood cells. A ventilation-perfusion lung scan was interpreted as showing a low probability for pulmonary embolus. A pulmonary artery catheter was placed, and his initial pulmonary capillary wedge pressure was 28 mm of mercury, which decreased to 18 mm of mercury within several hours. He had an elevated cardiac index (>7.5) and low systemic vascular pressures, and an echocardiogram documented normal ventricular function.

ABBREVIATIONS USED IN TEXT

ANA = antinuclear antibody
LDH = lactate dehydrogenase
SLE = systemic lupus erythematosus

A course of intravenously administered methylprednisolone sodium succinate, 125 mg every six hours, was started. He improved, and mechanical ventilation was discontinued after three days. Two days later, he became acutely hypoxemic and again required mechanical ventilation. A chest radiograph at that time revealed clearing of the left lung infiltrates and a new right upper lobe infiltrate. His hematocrit decreased abruptly from 0.27 to 0.23. He improved rapidly and was extubated two days later. Within a week, his lung infiltrates had completely resolved, he had no respiratory symptoms, and he had normal oxygen saturation while breathing room air. His regimen of methylprednisolone lasted a total of nine days: 500 mg per day for four days, 300 mg per day for four days, and 160 mg for one day; this was followed by a course of oral prednisone for four days. He required no further immunosuppressive therapy.

Ten months after the incident, the patient had no respiratory symptoms and had normal pulmonary function: forced vital capacity and forced expired volume in one second, both 100% of predicted values; total lung capacity, 99% of predicted values; and single-breath diffusing capacity, 88% of predicted values. His hematuria, hypertension, and proteinuria had completely resolved, and he had had no recurrence of skin rashes, arthralgia, or abdominal pain. He remains disease-free at the time of this report, more than 18 months since his initial presentation.

Discussion

Henoch-Schönlein purpura, a vasculitic syndrome of unknown cause, is usually a mild, self-limited disease most commonly affecting children between 2 and 10 years of age, but it has been observed in older children and adults.[2-4] Serious complications of the syndrome usually involve the kidneys (nephritis) and occasionally the gastrointestinal tract or the central nervous system. Clinically important pulmonary complications are rare.

Our patient had three of the four American College of Rheumatology criteria for Henoch-Schönlein purpura: palpable purpura without thrombocytopenia, age 20 years or younger at disease onset, and gastrointestinal bleeding.[5]

The fourth criterion, extravascular or perivascular granulocytes on skin biopsy, was not met because our patient did not have a biopsy. The differential diagnosis for this patient includes other vasculitic diseases that have been associated with pulmonary hemorrhage: Wegener granulomatosis, systemic lupus erythematosus (SLE), and polyarteritis nodosa. Our patient had a normal ANA level, indicating a less than 0.14% chance of having SLE.[6(p1039)] His complement levels were slightly decreased, which is consistent with SLE, but about 19% of patients with Henoch-Schönlein purpura also have low levels.[5] Wegener granulomatosis can be difficult to differentiate from Henoch-Schönlein purpura early in the disease course.[7] We did not assess for antineutrophilic cytoplasmic antibodies primarily because at the time of evaluation, the diagnosis of Henoch-Schönlein purpura was not in question. Nevertheless, we think that our patient did not have Wegener granulomatosis because he did not present with any upper respiratory tract clinical findings, and without any long-term immunosuppressive therapy, he has remained free of respiratory symptoms for more than a year after the onset of the illness. This would be extremely unusual for a patient with Wegener granulomatosis.[8]

Polyarteritis nodosa can present similarly to Henoch-Schönlein purpura. There are no specific laboratory tests that can confirm a diagnosis of polyarteritis nodosa, but histologic examination of involved tissues can be useful. We did not do any biopsies on our patient, but we think that, although we did not rule out polyarteritis nodosa, it was unlikely in our patient for the following reasons. It is rare in children. The rash of this disorder tends to persist for weeks and to extend over the entire body,[9] whereas our patient had the characteristic Henoch-Schönlein purpuric rash over the lower extremities and buttock, which resolved within two weeks. Polyarteritis nodosa is characterized by a progressive course with an extremely high mortality if not treated with long-term immunosuppressive therapy.[6(pp1238-1239),9,10] Our patient, after receiving just one short course of corticosteroids, has remained free of any skin, renal, or lung problems for more than a year since his acute illness began. Although possible, this would be highly unlikely if he had polyarteritis nodosa.[6]

Even though serious pulmonary complications of Henoch-Schönlein purpura are rare, they have been reported, with the most common one being pulmonary hemorrhage. We identified 15 cases of pulmonary hemorrhage associated with this disorder reported in the literature.[2-4,12-18] Of the 15 patients, 6 died as a result of the pulmonary hemorrhage, and all suffered serious morbidity. Of the 15 patients, 11 were either adolescents or adults, and all had severe, multiorgan involvement.

Although the vast majority of patients in whom Henoch-Schönlein purpura develops are 2 to 10 years old, most of those reported to have associated pulmonary hemorrhage have been adolescents and adults. This suggests that pulmonary hemorrhage is more common in adolescents and adults with this syndrome than it is in younger children.

The principal cause of pulmonary hemorrhage in Henoch-Schönlein purpura is a vasculitis of the small pulmonary blood vessels, especially the capillaries.[13,14,17] This patient's elevated pulmonary capillary wedge pressure might have increased his risk of pulmonary hemorrhage. In addition, he had mild uremia that could have affected platelet function, further predisposing him to hemorrhage. Corticosteroids have been used in the hope of ameliorating the vasculitis, but their efficacy has not been established. Both lack of response[14] and dramatic improvement[2] have been reported. Nevertheless, it seems prudent to use them in high doses (200 to 1,000 mg of methylprednisolone per day) until the event has resolved and then to taper.

Most of the patients reported in the literature with Henoch-Schönlein purpura and pulmonary hemorrhage were older and had more severe multiorgan involvement than the typical child with this syndrome. Patients with severe manifestations of Henoch-Schönlein purpura, especially adolescents and adults, are at risk of having acute, life-threatening pulmonary hemorrhage. Although not proved, early treatment of this high-risk group with corticosteroids should be considered in the hope of decreasing the morbidity and mortality of this serious complication.

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